

МІНІСТЕРСТВО ОХОРОНИ ЗДОРОВ'Я УКРАЇНИ
Харківський національний медичний університет

Module 3. Internal medicine.
Meaning module 1. Keeping patients
in cardiological clinic.
Theme 15. Management of patient
with a heart murmur

Guidelines for students

Модуль 3. Внутрішня медицина.
Змістовний модуль №1.
Ведення хворих у кардіології.
Тема 15. Ведення пацієнта з шумом у серці

Методичні вказівки
для студентів та лікарів-інтернів

Рекомендовано
вченою радою ХНМУ.
Протокол № 11 від 21.12.2011.

Харків
ХНМУ
2012

Module. Internal medicine. Contents module №1. Theme 15. Management of patient with a heart murmur / comp.: Oleg Babak, Galyna Fadeyenko, Olga Zaichenko et al. – Kharkiv: KNMU, 2012. – 36 p.

Compilers Oleg Babak,
 Galyna Fadeyenko.,
 Olga Zaichenko,
 A.B. Tveretinov
 Yu.N. Shaposhnikova

Модуль. Внутрішня медицина. Змістовний модуль №1. Тема 15. Ведення пацієнта з шумом у серці: метод вказ. для студентів та лікарів-інтернів / упор. О.Я. Бабак, Г.Д. Фадєєнко, О.Є. Зайченко та ін. – Харків: ХНМУ, 2012. – 36 с.

Упорядники Бабак О.Я.,
 Фадєєнко Г.Д.,
 Зайченко О.Є.
 Тверетінов О.Б.
 Шапошнікова Ю.М.

1. **Module title:** «Current practice of internal medicine».
2. **Theme title:** «Management the patient with a heart murmur».
3. **Theme specifications.**
4. **Theme aims.**

The aim of this module is to provide the student with an opportunity to keeping patient in cardiology clinic.

5. Learning outcomes

Student must know causes of heart murmurs. Etiology, pathogenesis. Haemodynamics. Clinical finding, treatment. Surgical treatment the Heart valve disease.

Students should be able

1. To execute assessment, diagnosis and management the patient with a heart murmur.
2. To obtain the skills of interpretation of Doppler echocardiography.
3. To obtain the skills of interpretation of the results of stress tests.
4. To obtain the skills of interpretation of electrocardiograms.

6. TESTS AND ASSIGNMENTS FOR SELF-ASSESSMENT

BASIC LEVEL OF KNOWLEDGE: MULTIPLE CHOICE QUESTIONS (CHOOSE THE CORRECT ANSWER/STATEMENT)

1. A female rheumatic patient experiences diastolic thoracic wall tremor (diastolic thrill), accentuated S1 at apex, there is diastolic murmur with presystolic intensification, opening snap, S2 accent at pulmonary artery. What kind of heart disorder is observed?

- | | |
|---------------------------------------|--------------------------------------|
| <i>A. Aortic valve insufficiency.</i> | <i>D. Opened arterial duct.</i> |
| <i>B. Mitral stenosis.</i> | <i>E. Pulmonary artery stenosis.</i> |
| <i>C. Mitral valve insufficiency.</i> | |

2. All of the following clinical findings are consistent with severe mitral stenosis **except**

- | | |
|---|------------------------------------|
| <i>A. Atrial fibrillation.</i> | <i>D. Pulsatile liver.</i> |
| <i>B. Opening snap late after S2.</i> | <i>E. Right-ventricular heave.</i> |
| <i>C. Pulmonary vascular congestion</i> | |

3. The most frequent type of valvular heart disease in the elderly is:

- | | |
|---------------------------------|---------------------------------|
| <i>A. Pulmonary stenosis.</i> | <i>D. Aortic stenosis.</i> |
| <i>B. Mitral stenosis.</i> | <i>E. Aortic regurgitation.</i> |
| <i>C. Mitral regurgitation.</i> | |

4. After objective clinical examination a 18 year old male was diagnosed with mitral valve prolapse. What complementary instrumental method of examination should be applied for the diagnosis confirmation?

A. *Phonocardiograph.*

D. *Echocardiography.*

B. *ECG.*

E. *Veloergometry.*

C. *Roentgenography of chest.*

5. A 42 y.o. woman complains of dyspnea, edema of the legs, and tachycardia during small physical exertion. Heart borders are displaced to the left and SI is accentuated, there is diastolic murmur on apex. The liver is enlarged by 5 cm. What is the cause of heart failure?

A. *Mitral stenosis.*

D. *Mitral regurgitation.*

B. *Tricuspid stenosis.*

E. *Tricuspid regurgitation.*

C. *Aortic stenosis.*

7. Specification of the theoretical question for training of «Management the patient with a heart murmur»

1. Mitral stenosis. Etiology, pathogenesis. Hemodynamics. Clinical finding, treatment. Surgical treatment.

2. Mitral regurgitation. Etiology, pathogenesis. Hemodynamics. Clinical finding, treatment. Surgical treatment.

3. Aortic valve stenosis. Etiology, pathogenesis. Hemodynamics. Clinical finding, treatment. Surgical treatment.

4. Aortic insufficiency. Etiology, pathogenesis. Hemodynamics. Clinical finding, treatment. Surgical treatment.

5. Tricuspid regurgitation. Etiology, pathogenesis. Hemodynamics. Clinical finding, treatment. Surgical treatment.

6. Tricuspid stenosis. Etiology, pathogenesis. Hemodynamics. Clinical finding, treatment. Surgical treatment.

7. Indicative syllabus.

The patient with a heart murmur Genesis and types of heart murmurs

Murmurs are defined as sounds heard in addition to the sequence of two to three heart sounds during each heartbeat. The two normal heart sounds—the first heart sound (S_1) and the second heart sound (S_2)—are produced mainly by the closure of the atrioventricular (tricuspid and mitral) and semilunar (aortic and pulmonary) valves, respectively. Occasionally, an additional heart sound associated with the ventricles filling up with blood may be heard. It is referred to as S_3 when it occurs in early diastole and S_4 when heard in late diastole after the atrial contraction.

Heart murmurs can be systolic, diastolic or continuous and they can be reported with accompanying intensity grades. When clinically insignificant, murmurs are referred to as being "innocent"; they are caused by increased flow or turbulence across anatomically normal valves. Some systolic murmurs may be clinically innocent, while all diastolic and continuous murmurs are abnormal.

Causes

Innocent Heart Murmurs. Innocent murmurs are heard when blood moves noisily through a normal heart. Sometimes these murmurs occur when:

- Blood is flowing faster than usual through the heart and blood vessels attached to it.

- An increased amount of blood is flowing through the heart.

Illnesses or conditions that can cause blood to flow to be faster than usual through the heart include:

- Fever
- Anemia
- Hyperthyroidism
- Pregnancy.

Congenital heart murmurs. Congenital heart murmurs are heard in the newborn. They may be due to abnormalities in the valves, septae or arteries, and veins that carry blood to and from the heart. In some complicated heart disease conditions there may be a combination of all three. Many congenital heart murmurs resolve spontaneously without medical intervention while others require surgical operations for repair.

Common congenital heart defects include the following:

- **Atrial septal defect (ASD)** is a hole in the septum (wall) that divides the heart's upper chambers (atria).

- **Ventricular septal defect (VSD)** is a hole in the septum that divides the heart's lower chambers (ventricles). More than half of all abnormal heart murmurs in children are caused by ventricular septal defects.

- **Patent ductus arteriosus (PDA)** occurs when the ductus arteriosus does not close after birth. A developing fetus does not breathe while in the uterus; therefore, its lungs do not require a blood supply. Before birth, a channel called the ductus arteriosus diverts blood around the lungs by connecting the pulmonary arteries, which supply blood to the lungs after birth, to the aorta, which supplies blood to the rest of the body. After birth, when the infant is breathing and the lungs fill with air, the ductus arteriosus normally closes, allowing blood to flow through the pulmonary arteries to the lungs. In a baby with PDA, the channel stays open and blood continues to pass by the lungs.

- **Cardiomyopathy** causes the heart muscle to become too thick or too weak to pump blood normally.

• **Stenosis** occurs when heart valves are "sticky" or too narrow, affecting blood flow into or out of the heart.

• **Regurgitation** occurs when heart valves do not close completely and blood leaks backward into the heart.

In cyanotic lesions, a portion of nonoxygenated blood never reaches the lungs; instead, it is shunted to the systemic circulation. Examples include transposition of great arteries, tetralogy of Fallot and single ventricle. Certain people with congenital heart disease have a characteristic body habitus, such as patients with trisomy 21 (Down syndrome). They have an increased prevalence of atrial or ventricular septal defects and tetralogy of Fallot.

Furthermore, congenital heart disease is associated with heritable connective tissue disorders such as osteogenesis imperfecta, Ehlers-Danlos syndrome, Marfan syndrome and Stickler's syndrome.

In adults and older children, heart murmurs are typically caused by **heart valve problems** related to infections, illness, or aging. These conditions include the following:

• **Rheumatic fever** is an inflammatory illness that can develop when strep throat infection is not properly treated. Rheumatic fever may eventually damage heart valves permanently.

• **Endocarditis** is a bacterial infection that attacks the inner lining of the heart. Left untreated, it can damage or destroy heart valves, block blood flow, or cause blood to leak backward.

• **Mitral valve prolapse** occurs when the mitral valve does not close properly. The mitral valve is located between the left atrium and left ventricle. When this valve doesn't close as it should, the "leaflets" of the valve expand back into the atrium when the left ventricle contracts. Mitral valve prolapse sometimes is a congenital heart defect that is not discovered until adulthood.

Valve calcification may develop as people age. In this condition, the heart valves harden or thicken, making it more difficult for blood to move through them.

• **Cardiac myxoma**, which is the most common type of heart tumor in adults, can grow inside of the heart and block blood flow.

• **Hypertrophic cardiomyopathy** (also called idiopathic hypertrophic subaortic stenosis) is a condition that causes the muscle inside the left ventricle to thicken, narrowing the path for blood flow.

Worldwide, the most common cause of clinically significant heart murmurs is rheumatic heart disease (RHD). In the United States and Canada, however, RHD is an uncommon finding except among recent immigrants. The most common causes of clinically significant murmurs in this country are degenerative valvular disorders such as senile calcific aortic stenosis (AS) and mitral valve prolapse (MVP). Less common causes of

acquired valvular disease are systemic lupus erythematosus (SLE) and certain medications used in weight loss programs.

Rheumatic heart disease. RHD is triggered by group A streptococcal pharyngitis. The microorganism induces an autoimmune reaction that may lead to valvular scarring and calcifications, especially involving the mitral valve. The most common forms of RHD are mitral stenosis and aortic insufficiency, both of which produce diastolic murmurs.

Aortic stenosis. Owing to aging, an initially normal trileaflet aortic valve may undergo calcific degeneration. Subsequent narrowing of the aortic valve orifice results in the clinical manifestations of AS in the elderly. When calcific aortic stenosis is encountered in middle-aged people, the clinician should suspect a diagnosis of congenitally bicuspid aortic valve (BAV). BAV is the most common form of congenital heart disease, occurring in 1 to 2 percent of all live births.

Mitral valve prolapse. Myxomatous degeneration of valve leaflets, typically those of the mitral valve, results in leaflet redundancy and systolic billowing of one or both leaflets into the left atrium. Sometimes, this is accompanied by regurgitation. Although the prognosis for MVP is good in general, it is the valvular disease that necessitates the most valve surgery in the United States. In cases in which MVP is associated with regurgitation, a murmur often will be heard preceded by a systolic click. Antibiotic prophylaxis is warranted for patients with MVP only when mitral regurgitation is present.

Systemic lupus erythematosus. Verrucous valvular lesions in SLE can be found in up to 60 percent of affected patients. Libman-Sacks endocarditis in patients with SLE refers to valvular thickening, followed by vegetations (that is, thrombi adherent to a diseased valve and composed of platelets, fibrin and sometimes microorganisms) and valvular insufficiency. Initially, the damage is caused by immune complex deposition on the valve. Patients with SLE who have Libman-Sacks endocarditis are at moderate risk of developing IE.

According to the guidelines of the American College of Cardiologists/American Heart Association, these patients should receive prophylactic antibiotics

Endocrine conditions. Patients with active or treated acromegaly have a high prevalence of mitral and aortic valve abnormalities characterized by fibrosis and/or calcifications of valve annulus and leaflets, which may lead to valvular regurgitation or stenosis.

All of the above-mentioned forms of acquired valvular disease confer a moderate risk of developing IE after dental procedures. Some forms of acquired valvular disease, however, carry a much higher risk. These include a history of IE and valve surgery.

DETECTION OF A HEART MURMUR

There are several ways may describe a murmur:

- Murmurs are classified ("graded") depending on how loud the murmur sounds with a stethoscope. The grading is on a scale. Grade I can barely be heard. An example of a murmur description is a "grade II/VI murmur." (This means the murmur is grade 2 on a scale of 1-6).

- Murmurs may be described as blowing, whooshing, or rasping.

- In addition, a murmur is described by the stage of the heartbeat when the murmur is heard. A heart murmur may be described as systolic or diastolic.

The following are important clues to the cause of the murmur:

- Does the murmur occur in the resting stage (diastole) or contracting stage (systole)?

- Does it occur early or late in the stage?

- Does it last throughout the heartbeat?

- Does it change when the doctor uses physical maneuvers?

- Can the murmur be heard in other parts of the chest, on the back, or in the neck?

- Where is the murmur heard the loudest?

Auscultation. The most common abnormal auscultatory finding on cardiac examination is a *systolic murmur*, which occurs in 80 to 96 percent of children and in 15 to 44 percent of adults. These murmurs can be functional (innocent) or pathological.

The current American College of Cardiologists/American Heart Association guidelines for the treatment of patients with valvular heart disease state that cardiac auscultation remains the most widely used technique of screening for heart disease, and that a careful observer will be able to deduce the correct origin and significance of a heart murmur. The physician should use echocardiography to confirm an impression or diagnosis of structural heart disease.

Echocardiography. Echocardiography is a diagnostic test that uses ultrasound waves to make images of the heart chambers, valves and surrounding structures. The purpose of echocardiography with regard to heart murmurs is to evaluate the primary lesion in terms of cause and severity, define the lesion's hemodynamic significance, detect coexisting abnormalities, evaluate the size and function of cardiac chambers and establish a reference point for future comparisons.

The standard echocardiography is called *transthoracic echocardiography* (TTE). In this procedure, the clinician performs cardiac imaging after placing an ultrasound probe on various points on the external chest and abdominal walls.

During transesophageal echocardiography (TEE), the heart and large vessels are imaged after a small ultrasound probe is introduced into the esophagus.

gus and stomach. It is considered an invasive procedure and should be used only when information beyond that obtained from conventional TTE is needed.

Overall, TTE has a lower sensitivity (60 to 70 percent) than does TEE (75 to 95 percent) in detecting infective endocarditis, particularly that of left-sided (mitral and aortic) valves. However, for the detection of infective endocarditis of the tricuspid valve, TTE often is as sensitive as TEE.

Both TTE and TEE are capable of M-mode, two-dimensional and Doppler imaging. Once echocardiographic images are acquired and stored on a videotape or computer disk, the cardiologist analyzes the examination findings and writes a report. The major components of this report are discussed below.

In view of the current literature, echocardiography is the procedure of choice for evaluating a heart murmur, but this technology should not be used as a "fishing net." Echocardiography probably is not a cost-effective method of screening asymptomatic patients for valvular heart disease. It should be reserved for dental patients with a moderate-to-severe risk of developing IE. When the clinician suspects IE, a positive echocardiographic finding is one of several major Duke criteria for the establishment of an IE diagnosis

Echocardiographic report. The echocardiographic report has many components. The normal values may vary from one medical center to another, but the various components of a standard report are similar. As with all laboratory reports, the initial review should make sure that the patient's name on the report is correct. The date and type of study (TTE or TEE) follow, along with general information about the patient (for example, age, sex, location of care).

The information regarding chamber sizes and wall thickness often is obtained via M-mode echocardiography. A descriptive section of the report follows the numeric data in which specific cardiac pathology (if present) is mentioned, and its hemodynamic significance is discussed based on the data from all echocardiographic imaging modalities.

One of the *most important parameters of cardiac function is the left ventricular ejection function*. An ejection fraction below 50 percent suggests left ventricular systolic dysfunction (that is, decreased left ventricular contractility).

GENERAL MEDICAL TREATMENT OF PATIENTS WITH MURMURS

The clinician's approach to treating patients with heart murmurs depends on many variables, including the *characteristics of the murmur itself (such as the intensity and type of murmur) and the presence or absence of cardiac symptoms*.

For example, an extensive work-up is not necessary for patients with grade 1 and 2 midsystolic murmurs if they are children or young adults in

whom the cardiac examination results are normal, and who do not have other signs and symptoms associated with cardiac disease.

However, if patients have signs and symptoms indicating the presence of IE, thromboembolism, congestive heart failure, myocardial infarction or syncope, echocardiography is indicated.

The classic symptoms of cardiac disease include chest pain or chest discomfort, dyspnea, palpitations and syncope. In the review of systems, clinicians must ask these key questions, the answers to which will give them clues regarding whether the murmur is innocent or clinically significant.

As noted above, mild regurgitation of the mitral, tricuspid or pulmonic valves frequently is detected in asymptomatic patients, often in the absence of an audible murmur on auscultation. In principle, such patients do not require routine antibiotic prophylaxis before dental and other bacteremia-inducing medical procedures.

In the work-up of a patient with a pathological heart murmur, echocardiography is mandatory to confirm the diagnosis, establish the severity of the lesion and assess ventricular function and size.

All health professionals, including dental professionals, should educate their patients who have clinically significant heart murmurs about antibiotic prophylaxis, while cardiologists and other physicians should counsel patients about functional limitation, exercise and work restrictions, and pregnancy issues.

Mitral stenosis

Mitral stenosis (MS) is a narrowing of the inlet valve into the left ventricle that prevents proper opening during diastolic filling. Patients with mitral stenosis typically have mitral valve leaflets that are thickened, commissures that are fused, and/or chordae tendineae that are thickened and shortened

The normal mitral valve area is 4.0 to 5.0 cm². Narrowing of the valve area to <2.5 cm² must occur before development of symptoms.

A mitral valve area >1.5 cm² usually does not produce symptoms at rest. However, if there is an increase in transmitral flow or a decrease in the diastolic filling period, there will be a rise in left atrial pressure and development of symptoms. Thus, the first symptoms of dyspnea in patients with mild MS are usually precipitated by exercise, emotional stress, infection, pregnancy, or atrial fibrillation with a rapid ventricular response.

Etiology. Chronic rheumatic heart disease is much the commonest cause of mitral stenosis, though there are a number of other well defined conditions in which blood flow across the mitral valve is limited to a variable extent.

1. Congenital mitral stenosis is a rare condition with thick, rolled leaflets and short chordae, with the spaces between them obliterated. The papillary muscles may be abnormally inserted, either directly from the free wall of the ventricle or from the septum. In parachute mitral valve, there is only one papillary muscle.

Congenital mitral stenosis may be associated with left ventricular outflow obstruction, hypoplasia of the left ventricular cavity and the aorta, or endocardial fibroelastosis.

2. A calcified mitral valve ring may rarely cause mild mitral stenosis.

3. In infective endocarditis, bulky vegetations may occasionally interfere with transmitral flow.

4. Nodular rheumatoid arthritis may be associated with thickening of the valve cusps, but true mitral stenosis does not occur.

5. In systemic lupus erythematosus, treatment of Libman–Sachs endocarditis with steroids has led to fibrosis of the cusps with commissural fusion.

6. The combination of ostium secundum atrial septal defect and rheumatic mitral stenosis, Lutembacher syndrome, is probably fortuitous.

Rheumatic mitral stenosis

Incidence. The incidence of rheumatic mitral stenosis parallels that of acute rheumatic fever. It is thus much commoner, and presents earlier, in the Middle East, the Indian sub-continent, and the Far East than in the West.

Clinical features

History. Inquire about the history of acute rheumatic fever (RF), although 50–60% of patients do not recall this. Also inquire about history of repeated streptococcal pharyngitis or scarlet fever in childhood.

Many patients are asymptomatic. Some patients may develop.

- Symptoms during physiologic stress such as infection, exercise, fever, or pregnancy.

- Exertional dyspnea, orthopnea, and paroxysmal nocturnal dyspnea (symptoms of left heart failure) are most common. Dyspnea may be accompanied by cough and wheezing. Attacks of frank pulmonary edema may occur.

- Chest pain due to right ventricular ischemia, concomitant coronary atherosclerosis, or a coronary embolism may be present.

- Hemoptysis from pulmonary venous hypertension results in rupture of anastomosis between bronchial veins.

- Ortner syndrome may occur if an enlarged left atrium impinges on the left recurrent laryngeal nerve, causing hoarseness.

- Patients may present with complications of mitral stenosis.

- New-onset atrial fibrillation.

- Systemic embolism.

- Infective endocarditis.

Physical. Look for findings not only intrinsic to valvular deformity but also hemodynamic disturbance from the stenotic valve and its complications.

Symptoms

- Cardiac examination of stenotic mitral valve (best at the apex with the patient in the left lateral recumbent position).

- Palpable diastolic thrill
- An accentuated S₁, followed by S₂, and an opening snap (OS)
- Characteristic diastolic low-pitched, rumbling murmur
- The duration, and not intensity, of the murmur is a guide to the severity of mitral valve narrowing. However, murmur may diminish in intensity as the stenosis increases.

- The OS and diastolic murmur are often reduced during inspiration and augmented during expiration. Amyl nitrite inhalation, coughing, isometric or isotonic exercise, and sudden squatting all are useful in accentuating a faint or equivocal murmur of mitral stenosis.

- Signs of left heart failure.

- Respiratory distress, evidence of pulmonary edema (eg, rales)

- Digital clubbing.

- Signs of right heart strain/failure.

- Right ventricular lift may be felt. A loud pulmonic closure (P₂) may be noted in the left parasternal region in patients with pulmonary hypertension.

- Jugular venous distention, ascites, hepatomegaly, and peripheral edema may be noted.

- Auscultation may reveal a systolic murmur of TR, a Graham Steele murmur of PR (a high-pitched, decrescendo, diastolic murmur of pulmonary insufficiency), and an S₄.

- Signs of complications from mitral stenosis.

- Endocarditis - Fever, murmur, and classically splinter hemorrhage, petechiae, Roth spots, Osler nodes, or Janeway lesions.

- Atrial fibrillation

- Systemic embolization

- Other findings.

- A holosystolic murmur of mitral regurgitation may accompany the valvular deformity of mitral stenosis.

- "Mitral facies" characterized by pinkish purple patches on the cheeks may be present.

Investigations

Chest radiography

- Look for left atrial, pulmonary artery, right ventricle, and/or right atrium enlargement (eg, straightening of left heart border, loss of aortic window).

- Rarely, calcification of the mitral valve may be seen.

- Radiologic changes in the lung fields indirectly reflect the severity of mitral stenosis.

- Interstitial edema manifests as Kerley B lines.

- Severe, long-standing mitral obstruction results in Kerley A lines and findings of pulmonary hemosiderosis.

- Pulmonary edema is seldom evident on the chest radiography.

Echocardiography

- Two-dimensional (2D) and Doppler echocardiography (echo) is the diagnostic study of choice. It should be performed in order to make the diagnosis and to assess valve function whenever a change in symptoms or physical examination findings is noted.

- 2D echocardiography evaluates the morphology of the mitral valve, measuring orifice size and detailing leaflet mobility, thickness, calcification, fusion, and appearance of the commissures. It provides anatomic and functional information on cardiac chambers and facilitates recognition of other structural abnormalities.

- **Doppler echocardiography** is the most accurate noninvasive technique to quantify the hemodynamic severity of mitral stenosis at rest or with exercise. It measures transvalvular pressure gradient and pulmonary arterial pressure and determines whether mitral regurgitation, aortic regurgitation, and other valvular abnormalities coexist.

- If a question exists about the diagnosis after transthoracic echocardiography, a transesophageal echocardiography (TEE) provides better images of the mitral valve and is a more sensitive way to detect pathology such as valvular vegetations or atrial thrombus.

Cardiac catheterization. This is rarely necessary, either to make the diagnosis or to assess severity. It is performed only to determine the state of the coronary arteries in older patients and as a prelude to balloon valvuloplasty, or very occasionally in patients in whom diagnostic echocardiograms cannot be obtained

Other Tests

- **ECG** is relatively insensitive for mild mitral stenosis.

- Ninety percent of patients with significant mitral stenosis and sinus rhythm display electrical evidence of left atrial enlargement: P-mitrale in lead II and/or a biphasic P wave in lead V₁ with a wide negative deflection greater than 0.04 seconds.

- The QRS axis in the frontal plane correlates with the severity of valve obstruction in pure mitral stenosis. A mean axis between 0 and +60 degrees suggests a mitral valve area $>1.3 \text{ cm}^2$, whereas an axis of more than 60 degrees suggests a valve area $<1.3 \text{ cm}^2$.

- Atrial fibrillation usually develops in the presence of preexisting left atrial enlargement.

- With severe pulmonary hypertension, right-axis deviation and right ventricular hypertrophy can be seen. The ECG of right ventricular hypertrophy

typically shows tall R waves in the right chest leads, and the R wave may be taller than the S wave in lead V₁. In addition, right-axis deviation and right precordial T-wave inversions are often present.

- **Exercise stress testing**

- Exercise stress testing is indicated in situations where the degree of disability is in question.

- Stress echocardiography will provide information about changes in the transmitral gradient and the degree of limitation of exercise and may guide decisions about valvotomy.

- **Laboratory Studies**

- Ruling out other diseases is useful.

- Brain natriuretic peptide may be useful in determining the presence of heart failure in an undifferentiated patient with dyspnea.

- Troponin I and creatinine kinase levels may be useful in ruling out acute myocardial infarction in patients who present with symptomatic mitral stenosis.

Diagnosis. The diagnosis of mitral stenosis is usually straightforward on the basis of history, physical signs, and chest radiography, and can rapidly be confirmed by echocardiography. When the ventricular rate is rapid, the diastolic murmur may be inaudible, but becomes apparent when the ventricular rate is controlled by digoxin. Silent mitral stenosis may mimic primary pulmonary hypertension, but the correct diagnosis is easily made by echocardiography. Mild mitral stenosis should be suspected as a source of systemic emboli and as a cause of unexplained atrial fibrillation, particularly in the elderly.

- **Differential diagnosis**

1. Left atrial myxoma
2. cor triatriatum
3. pulmonary veno-occlusive disease or
4. Austin–Flint murmur

- **Treatment**

Emergency Department Care. The goal is to control symptoms, to prevent or retard disease progression, and to treat complications.

- Treatment of congestive heart failure

- Medications to consider include nitroglycerin, ACE inhibitors, and diuretics.

- Patients with severe mitral stenosis should maintain an upright posture and avoid strenuous physical activity.

- Sodium intake should be restricted, and maintenance doses of oral diuretics should be continued.

- The data on beta-blockers are conflicting; beta-blockade may be useful for patients with exertional symptoms if the symptoms occur primarily at high heart rates.

- Prevent or retard disease. Primary and/or secondary prophylaxis against streptococci/endocarditis should be administered.

- Penicillin is indicated whenever streptococcal infection is suspected in a patient with known rheumatic disease.

- Management of atrial fibrillation

- Much of the dyspnea related to mitral stenosis is rate related. Control of atrial fibrillation with rapid ventricular response may be considered with any of the following agents:

- ♦ If the patient is unstable and immediate cardioversion is indicated (synchronized 100 J, 200 J, 300 J, then 360 J monophasic, or biphasic equivalent, with prior sedation), then heparin should be administered before, during, and after cardioversion. Otherwise, electrical or chemical cardioversion should be performed after 3 weeks of warfarin anticoagulation.

- ♦ Anticoagulation is necessary in patients who are unable to maintain normal sinus rhythm.

- ♦ Anticoagulation may also be beneficial for patients with normal sinus rhythm with a prior embolic event or a left atrial dimension greater than 55 mm Hg noted by echocardiography.

Consultations

- Cardiologist and/or cardiac thoracic surgeon

- Known or suspected cases of mitral stenosis with hemodynamic instability, arrhythmia, or embolization.

- Cases involving a new onset or progression of symptoms.

Medication. The goal of medical therapy is to control the rapid ventricular rate and to prevent thrombus formation and embolization. Conversion of atrial fibrillation to sinus rhythm may also decrease symptoms.

Antiarrhythmics. These agents alter the electrophysiologic mechanisms that are responsible for arrhythmia.

• Amiodarone (Cordarone)

Class III antiarrhythmic. Has antiarrhythmic effects that overlap all 4 Vaughn-Williams antiarrhythmic classes. May inhibit AV conduction and sinus node function. Prolongs action potential and refractory period in myocardium and inhibits adrenergic stimulation. Only agent proven to reduce incidence and risk of cardiac sudden death, with or without obstruction to LV outflow. Very efficacious in converting atrial fibrillation and flutter to sinus rhythm and in suppressing recurrence of these arrhythmias.

Has low risk of proarrhythmia effects, and any proarrhythmic reactions generally are delayed. Used in patients with structural heart disease. Most clinicians are comfortable with inpatient or outpatient loading with 400 mg PO tid for 1 wk because of low proarrhythmic effect, followed by weekly reductions

with goal of lowest dose with desired therapeutic benefit (usual maintenance dose for AF 200 mg/d). During loading, patients must be monitored for bradyarrhythmias. Prior to administration, control the ventricular rate and CHF (if present) with digoxin or calcium channel blockers.

Oral efficacy may take weeks. With exception of disorders of prolonged repolarization (eg, LQTS), may be DOC for life-threatening ventricular arrhythmias refractory to beta-blockade and initial therapy with other agents.

Adult. 120 mg IV over 10 min follow by IV infusion Maintenance dosing: 400 mg PO tid for 1-3 wk until desired effect achieved then followed by weekly reductions (goal of lowest dose with desired therapeutic benefit) Maintenance for AF: 200 mg/d PO

•**Digoxin (Lanoxin).** Cardiac glycoside that has direct inotropic effects in addition to indirect effects on the cardiovascular system. Effects on myocardium involve a direct action on the cardiac muscle that increases myocardial systolic contractions as well as indirect actions that result in increased carotid sinus nerve activity and enhanced sympathetic withdrawal for any given increase in mean arterial pressure.

Adult. 0.25 mg IV; up to 1 mg loading dose followed by a maintenance dose of 0.125-0.25 mg qd

•**Beta-adrenergic blockers.** These drugs inhibit chronotropic, inotropic, and vasodilatory responses to beta-adrenergic stimulation.

Metoprolol (Lopressor)

Selective beta1-adrenergic receptor blocker that decreases the automaticity of contractions. During IV administration, carefully monitor blood pressure, heart rate, and ECG.

Adult. 5 mg IV; repeat in 10 min; not to exceed 15 mg

•**Calcium channel blockers**

In specialized conducting and automatic cells in the heart, calcium is involved in the generation of the action potential. The calcium channel blockers inhibit movement of calcium ions across the cell membrane, depressing both impulse formation (automaticity) and conduction velocity.

Diltiazem (Cardizem CD, Cardizem SR, Tiazac, Dilacor). During the depolarization, inhibits the calcium ion from entering the slow channels or the voltage-sensitive areas of the vascular smooth muscle and myocardium.

Adult. 0.25 mg/kg IV over 2 min

Rebolus after 15 min prn with 0.35 mg/kg IV

•**Anticoagulants.** These agents inhibit thrombogenesis.

Heparin. Augments activity of antithrombin III and prevents conversion of fibrinogen to fibrin. Does not actively lyse but is able to inhibit further thrombogenesis. Prevents reaccumulation of clot after spontaneous fibrinolysis.

Most data are related to use of unfractionated heparin. Low molecular weight heparin probably is as effective but awaits the results from clinical studies.

Adult. 80U/kg IV initially, followed by a maintenance infusion of 18 U/kg/h IV; target aPTT is 50-70 s

Warfarin (Coumadin). Inhibits vitamin K-dependent clotting factors II, VII, IX, and X and anticoagulant proteins C and S. Anticoagulation effect occurs 24 h after drug administration, but peak effect may happen 72-96 h later. Antidotes are vitamin K and FFP.

Adult. 5-15 mg PO qd for 2-5 d, adjust to desired INR or PT; 2-10 mg/d PO qd maintenance Adjust dose to maintain INR of 2.5-3.5 or PT of 1.5-2 times baseline

•Further Inpatient Care

•Balloon valvotomy is, in general, the initial procedure of choice for symptomatic patients with moderate-to-severe mitral stenosis. It can double the mean valve area with a 50-60% decrease in transmitral gradient, producing a prominent and sustained symptomatic improvement.

▪ The 2006 ACC/AHA Practice Guidelines recommend percutaneous mitral balloon valvotomy for symptomatic patients (New York Heart Association Functional Class, or NYHA FC, II, III, or IV) with moderate or severe mitral stenosis (mitral valve area $\leq 1.5 \text{ cm}^2$), and favorable valve morphology in the absence of left atrial thrombus or significant mitral regurgitation.¹ Significant pulmonary artery hypertension (2/3 systolic blood pressure) is also an indication for intervention.

▪ In patients with indications for intervention, it has proven superior to closed commissurotomy in some long-term studies. The overall event-free (from death, repeat valvotomy, or valve replacement) survival rate is 80-90% in patients with favorable valve morphology. More than 90% of patients free of events remain in NYHA FC I or II. In general, surgical commissurotomy, either open or closed, has

•similar efficacy to balloon valvotomy. In the United States, open commissurotomy is considered the procedure of choice.

▪ The 2006 ACC/AHA Practice Guidelines gave a class I recommendation for surgical intervention in patients with NYHA FC III-IV symptoms, moderate or severe mitral stenosis, and valve morphology favorable for repair or replacement if one of the following exists:¹

- ♦ Percutaneous mitral balloon valvotomy is not available
- ♦ A left atrial thrombus is present despite anticoagulation
- ♦ A nonpliable or calcified valve with the decision to proceed with either repair or replacement made at the time of the operation

▪ Patients with moderate or severe mitral regurgitation should undergo mitral valve replacement.

- The maze procedure may be useful to correct concomitant atrial fibrillation in patients undergoing valve replacement or surgical commissurotomy.

Inpatient & Outpatient Medications

Medications cannot correct mitral stenosis, rather therapies are directed at reducing the incidence and severity of symptoms and complications.

- Drugs that increase diastolic filling time and decrease the heart rate are typically used.

- Beta-blockers are frequently used in this situation

- Calcium channel blockers, such as diltiazem, and digoxin may also be used. Beta-blockers are preferred over digoxin because they control exercise-induced increases in heart rate.

- Rhythm control is of questionable clinical significance. Amiodarone may be used to maintain sinus rhythm, but its use may cause complications.

- Anticoagulation is used in patients with atrial thrombi, in patients with atrial fibrillation, or in patients with a prior thromboembolic event.

Deterrence/Prevention

- Primary prophylaxis consists of an early diagnosis of group A streptococcal pharyngitis. Treatment started within 7-9 days after onset of illness may prevent rheumatic fever. Treatment with penicillin or if allergic (a macrolide) is recommended. To date, no strains of streptococci that produce rheumatic fever have been penicillin resistant.

- The prevention of repeated attacks may delay the progression of mitral stenosis. Secondary prophylaxis may be individually tailored.

- With carditis, secondary prevention continues for 10 years or until age 25 years.

- Without carditis, secondary prevention continues for 5 years or until age 18 years.

- Endocarditis prophylaxis for routine dental and respiratory procedures is no longer recommended for patients with mitral stenosis unless they have had implantation of an artificial valve.

Complication

- Thromboembolism.

- Atrial fibrillation.

- Bacterial endocarditis.

- Pulmonary hypertension.

- Pulmonary edema.

- Complications of balloon valvulotomy (eg, stroke, cardiac perforation, development of mitral regurgitation).

- Complications of mitral valve replacement (eg, paravalvular leakage, thromboembolism, infective endocarditis, mechanical dysfunction, bleeding due to anticoagulants).

Prognosis

• Depending on severity of symptoms of mitral stenosis (by the NYHA functional class), the 10-year survival rate is as follows:

- 85% for no symptom (class I).
- 34-42% for mild symptoms (early class II).
- 40% for moderate-severe symptoms (late class II, class III).
- 0% for class IV (Of class IV patients, survival is 42% at 1 year and 10% at 5 years.).

• The operative mortality rate is 1-2% for mitral commissurotomy and 2-5% for mitral valve replacement.

Mitral Regurgitation. Mitral regurgitation (MR) is incompetency of the mitral valve causing flow from the left ventricle (LV) into the left atrium during systole. Common causes include mitral valve prolapse, ischemic papillary muscle dysfunction, rheumatic fever, and annular dilation secondary to LV systolic dysfunction and dilation. Complications include progressive heart failure, arrhythmias, and endocarditis. Symptoms and signs include palpitations, dyspnea, and a holosystolic apical murmur. Diagnosis is by physical examination and echocardiography. Prognosis depends on LV function and severity and duration of MR. Patients with mild, asymptomatic MR may be monitored, but progressive or symptomatic MR requires mitral valve repair or replacement.

Etiology. MR may be acute or chronic. *Causes of acute MR include* ischemic papillary muscle dysfunction or rupture; infective endocarditis; acute rheumatic fever; spontaneous, traumatic, or ischemic tears or rupture of the mitral valve leaflets or subvalvular apparatus; acute dilation of the LV due to myocarditis or ischemia; and mechanical failure of a prosthetic mitral valve.

Common causes of chronic MR include those of acute MR plus myxomatous degeneration of the mitral leaflets or chordae tendineae, mitral valve prolapse (MVP), mitral annular enlargement, and nonischemic papillary muscle dysfunction (eg, due to LV enlargement). Uncommon causes of chronic MR include a congenital endocardial cushion defect with a cleft anterior leaflet, SLE, acromegaly, myxoma involving the valve or chordae, and calcification of the mitral annulus (mainly in elderly women).

Symptoms and Signs. Acute MR causes the same symptoms and signs as acute heart failure and cardiogenic shock. Most patients with chronic MR are initially asymptomatic and develop symptoms insidiously as the LA enlarges, pulmonary artery and venous pressure increases, and LV remodeling occurs. Symptoms include dyspnea, fatigue (due to heart failure), orthopnea, and palpitations (often due to AF). Rarely, patients present with endocarditis (eg, fever, weight loss, embolic phenomena).

Signs develop only when MR becomes moderate to severe. Inspection and palpation may detect a brisk apical impulse and sustained left parasternal movement due to systolic expansion of an enlarged LA. An LV impulse that is sustained, enlarged, and displaced downward and to the left suggests LV hypertrophy and dilation. A diffuse precordial lift occurs with severe MR because the LA enlarges, causing anterior cardiac displacement, and pulmonary hypertension causes right ventricular hypertrophy. A regurgitant murmur (or thrill) may also be palpable in severe cases.

- On auscultation, the 1st heart sound (S_1) may be soft (or occasionally loud). A 3rd heart sound (S_3) at the apex reflects a dilated LV and important MR.

The cardinal sign of MR is a holosystolic (pansystolic) murmur, heard best at the apex with the diaphragm of the stethoscope when the patient is in the left lateral decubitus position. In mild MR, the systolic murmur may be abbreviated or occur late in systole. The murmur begins with S_1 in conditions causing leaflet incompetency throughout systole, but it often begins after S_1 (eg, when chamber dilation during systole distorts the valve apparatus or when myocardial ischemia or fibrosis alters dynamics). When the murmur begins after S_1 , it always continues to the 2nd heart sound (S_2). The murmur radiates toward the left axilla; intensity may remain the same or vary. If intensity varies, the murmur tends to crescendo in volume up to S_2 . MR murmurs increase in intensity with handgrip or squatting because peripheral vascular resistance to ventricular ejection increases, augmenting regurgitation into the LA; murmurs decrease in intensity

- with standing or the Valsalva maneuver. A short rumbling mid-diastolic inflow murmur due to torrential mitral diastolic flow may be heard following an S_3 . In patients with posterior leaflet prolapse, the murmur may be coarse and radiate to the upper sternum, mimicking aortic stenosis. MR murmurs may be confused with tricuspid regurgitation, which can be distinguished because its murmur is augmented during inspiration.

Diagnosis

• Echocardiography

Diagnosis is suspected clinically and confirmed by echocardiography. Doppler echocardiography is used to detect regurgitant flow and help quantify its severity; 2-dimensional echocardiography is used to determine the cause of MR and to detect pulmonary hypertension.

If endocarditis or valvular thrombi are suspected, transesophageal echocardiography (TEE) can provide a more detailed view of the mitral valve and LA. TEE is also indicated when mitral valve repair instead of replacement is being considered to confirm the anatomy in more detail.

An ECG and chest x-ray are usually obtained initially.

• **ECG** may show LA enlargement and LV hypertrophy with or without ischemia. Sinus rhythm is usually present when MR is acute because the atria have not had time to stretch and remodel.

• **Chest x-ray** in acute MR may show pulmonary edema; abnormalities in cardiac silhouette are not evident unless an underlying chronic disorder is also present. Chest x-ray in chronic MR may show LA and LV enlargement. It may also show pulmonary vascular congestion and pulmonary edema with heart failure.

Cardiac catheterization is done before surgery, mainly to determine whether coronary artery disease (CAD) is present. A prominent systolic *c-v* wave is seen on pulmonary artery occlusion pressure (pulmonary capillary wedge pressure) tracings during ventricular systole. Ventriculography can be used to quantify MR.

Differential diagnosis

1. Ventricular septal defect

2. Aortic valve disease—The ejection systolic murmur of aortic valve disease is frequently audible at the apex, where it may be louder than at the base, and have a slightly different quality. However, this is not an adequate basis for diagnosing additional mitral regurgitation and it is essential to establish that the timing of the murmur is pansystolic, either from its relation to the second heart sound, or in aortic regurgitation, from its relation to the start of the early diastolic murmur.

3. Tricuspid regurgitation—The pansystolic murmur of tricuspid regurgitation may be mistaken for that of mitral regurgitation, particularly when the right ventricle is greatly enlarged. The presence of tricuspid regurgitation can be suspected from an elevated venous pressure with systolic waves, and confirmed by Doppler echocardiography. In severe mitral regurgitation, however, additional tricuspid regurgitation may be present.

Prognosis. Prognosis varies by acuity and cause of MR. Once MR becomes severe, about 10% of asymptomatic patients become symptomatic per year thereafter. About 10% of patients with chronic MR caused by MVP require surgical intervention.

Treatment

• Mitral valve repair or replacement

Acute MR requires emergency mitral valve repair or replacement; patients with ischemic papillary muscle rupture may also require coronary revascularization. Pending surgery, nitroprusside or nitroglycerin infusion may be used to reduce afterload, thus improving forward stroke volume and reducing ventricular and regurgitant volume.

Definitive treatment of chronic MR is also mitral valve repair or replacement, but patients with asymptomatic or mild chronic MR and no pulmo-

nary hypertension or AF may do well with periodic monitoring. ACE inhibitors or angiotensin receptor blockers are used to decrease left ventricular preload and afterload. They are used in patients with moderate mitral insufficiency to delay dilation of the LV. Loop diuretics such as furosemide are helpful in patients with exertional or nocturnal dyspnea. Digoxin may reduce symptoms in patients with atrial fibrillation or those in whom valve surgery is not appropriate. The ideal timing for surgery is uncertain, but intervention before ventricular decompensation (defined as echocardiographic end-diastolic dimension > 70 mm, end-systolic dimension > 45 mm, and ejection fraction < 60%) improves outcomes and decreases the chance of worsening LV function. After decompensation, ventricular function becomes dependent on the afterload reduction of MR, and in about 50% of decompensated patients, valve replacement causes a markedly depressed ejection fraction. For patients with moderate MR and significant CAD, perioperative mortality rate is 1.5% with bypass surgery alone and 25% with concomitant valve replacement. If technically feasible, valve repair instead of replacement is preferred; perioperative mortality rate is 2 to 4% (compared with 5 to 10% for replacement), and long-term prognosis is good (80 to 94% survival rate at 5 to 10 yr, compared with 40 to 60% for replacement).

Antibiotic prophylaxis is no longer recommended except for patients who have had valve replacement. Anticoagulants are used to prevent thromboemboli in patients with heart failure or AF. Although severe MR without mitral stenosis or AF is less likely to be complicated by atrial thrombosis, most cardiologists still recommend anticoagulants.

Mitral valve prolapse (MVP). Mitral valve prolapse (MVP) is a billowing of mitral valve leaflets into the left atrium during systole.

The most common cause is idiopathic myxomatous degeneration. MVP is usually benign, but

complications include mitral regurgitation, endocarditis, valve rupture, and possibly thromboembolism.

MVP is usually asymptomatic, in the absence of important regurgitation, although there are reports that some patients experience chest pain, dyspnea, dizziness, and palpitations. Signs include a crisp mid-systolic click, followed by a late systolic murmur if regurgitation is present. Diagnosis is by physical examination and echocardiography. Prognosis is excellent in the absence of significant regurgitation, but chordal rupture and endocarditis may occur. No specific treatment is necessary unless mitral regurgitation is present.

MVP is common; prevalence is 1 to 3% in otherwise normal populations, depending on the echocardiographic criteria used. Women and men are affected equally; onset usually follows the adolescent growth spurt.

Symptoms and Signs. Most patients are asymptomatic. Some experience nonspecific symptoms (eg, chest pain, dyspnea, palpitations, dizziness,

near syncope, migraines, anxiety), thought to be due to poorly defined associated abnormalities in adrenergic signaling and sensitivity rather than to mitral valve pathology.

In about one third of patients, emotional stress precipitates palpitations, which may be a symptom of benign arrhythmias (atrial premature beats, paroxysmal atrial tachycardia, ventricular premature beats, complex ventricular ectopy).

Occasionally, patients present with MR. Rarely, patients present with endocarditis (eg, fever, weight loss, thromboembolic phenomena) or stroke. Sudden death occurs in < 1%, most often resulting from ruptured chordae tendineae and flail mitral valve leaflets. Death due to a fatal arrhythmia is rare.

Typically, MVP causes no visible or palpable cardiac signs. MVP alone causes a crisp mid-systolic click heard best with the diaphragm of the stethoscope over the left apex when the patient is in the left lateral decubitus position. MVP with MR causes a click with a late-systolic MR murmur. The click becomes audible or moves closer to the 1st heart sound (S_1) and becomes louder with maneuvers that decrease left ventricle (LV) size (eg, sitting, standing, Valsalva maneuver); the same maneuvers cause an MR murmur to appear or become louder and last longer. These effects occur because decreasing LV size causes papillary muscles and chordae tendineae to pull together more centrally beneath the valve, resulting in quicker, more forceful prolapse with earlier, more severe regurgitation. Conversely, squatting or isometric handgrip delays the S_1 click and shortens the MR murmur. The systolic click may be confused with the click of congenital aortic stenosis, which can be distinguished because it occurs very early in systole and does not move with postural or LV volume changes. Other findings include a systolic honk or whoop, thought to be caused by valvular leaflet vibration; these findings are usually transient and may vary with respiratory phase. An early diastolic opening snap caused by return of the prolapsed valve to its normal position is rarely heard.

Diagnosis

Echocardiography. Diagnosis is suggested clinically and confirmed by 2-dimensional echocardiography. Holosystolic displacement of ≥ 3 mm or late systolic displacement of ≥ 2 mm identifies 95% of patients with MVP; the percentage is slightly higher if echocardiography is done while the patient is standing. Thickened, redundant mitral valve leaflets and displacement of ≥ 5 mm are thought to indicate more extensive myxomatous degeneration and greater risk of endocarditis and MR. Holter monitoring and 12-lead ECG may be useful for documenting arrhythmias in patients with palpitations.

Prognosis. MVP is usually benign, but severe myxomatous degeneration of the valve can lead to MR. In patients with severe MR, incidence of LV or left atrium enlargement, arrhythmias (eg, AF), infective endocarditis, stroke, need for valve replacement, and death is about 2 to 4%/yr.

Treatment

- Usually none
- Sometimes β -blockers

Treatment of MR depends on severity and associated left atrial and LV changes.

Antibiotic prophylaxis against endocarditis is no longer recommended. Anticoagulants to prevent thromboembolism are recommended only for patients with AF or prior transient ischemic attack or stroke.

Aortic stenosis (AS). Aortic stenosis (AS) is narrowing of the aortic valve obstructing blood flow from the left ventricle to the ascending aorta during systole.

The aortic valve area must be reduced to one fourth its normal size before significant changes in the circulation occur. Because the normal adult valve orifice is ≈ 3.0 to 4.0 cm^2 , an area ≥ 0.75 to 1.0 cm^2 is usually not considered severe AS. In large patients, a valve area of 1.0 cm^2 may be severely stenotic, whereas a valve area of 0.7 cm^2 may be adequate for a smaller patient.

The committee used a variety of hemodynamic and natural history data to grade the degree of AS as mild (area $>1.5 \text{ cm}^2$), moderate (area >1.0 to 1.5 cm^2), or severe (area $\leq 1.0 \text{ cm}^2$). When stenosis is severe and cardiac output is normal, the mean transvalvular pressure gradient is generally $>50 \text{ mm Hg}$. Some patients with severe AS remain asymptomatic, whereas others with only moderate stenosis develop symptoms. Therapeutic decisions, particularly those related to corrective surgery, are based largely on the presence or absence of symptoms. Thus, the absolute valve area (or transvalvular pressure gradient) is not usually the primary determinant of the need for aortic valve replacement (AVR).

Causes

- congenital bicuspid valve,
- idiopathic degenerative sclerosis with calcification,
- rheumatic fever.

Symptoms and Signs. Congenital AS is usually asymptomatic until at least age 10 or 20 yr, when symptoms may begin to develop insidiously. In all forms, progressive untreated AS ultimately results in exertional syncope, angina, and dyspnea (SAD triad). Other symptoms and signs may include those of HF and arrhythmias, including ventricular fibrillation leading to sudden death.

Physical

- The most common signs of aortic stenosis include a pulse of small amplitude.
- Palpation reveals a laterally displaced apex reflecting the presence of left ventricular hypertrophy.

- A systolic thrill may be palpable at the base of the heart, in the jugular notch, and along the carotid arteries.

Crescendo-decrescendo systolic ejection murmur begins shortly after the first heart sound. The intensity increases toward midsystole, then decreases, and the murmur ends just before the second heart sound. It is generally a rough, low-pitched sound that is loudest at the base of the heart and most commonly is

- Appreciated in the second right intercostal space that radiates to the carotids. An ejection click may be auscultated. This is associated with bicuspid valves.

- An audible fourth heart sound indicates the presence of left ventricular hypertrophy in severe aortic stenosis. Once the left ventricle dilates and fails, a third heart sound may be audible.

- Pulsus parvus et tardus: This is an arterial pulse with a delayed and plateaued peak, decreased amplitude, and gradual downslope.

- A high-pitched, diastolic blowing murmur may be present if the patient has associated aortic regurgitation.

Diagnosis

•Echocardiography

Diagnosis is suspected clinically and confirmed by echocardiography. Two-dimensional transthoracic echocardiography is used to identify a stenotic aortic valve and possible causes, to quantify LV hypertrophy and degree of diastolic or systolic dysfunction, and to detect coexisting valvular heart disorders (aortic regurgitation, mitral valve disorders) and complications (eg, endocarditis). Doppler echocardiography is used to quantify degree of stenosis by measuring aortic valve area, jet velocity, and transvalvular systolic pressure gradient.

A valve area of 0.5 to 1.0 cm² or a mean gradient > 45 to 50 mm Hg represents severe stenosis. The gradient may be overestimated in aortic regurgitation and underestimated in LV systolic dysfunction. The rate of progression of AS from mild to severe is quite variable and does not necessarily proceed in a linear fashion.

Cardiac catheterization is necessary to determine whether coronary artery disease (CAD) is the cause of angina and, occasionally, to resolve differences between clinical and echocardiographic findings.

- **ECG** typically shows changes of LV hypertrophy with or without an ischemic ST- and T-wave pattern.

- **Chest x-ray** findings may include calcification of the aortic cusps (seen on the lateral projection or on fluoroscopy) and evidence of HF. Heart size may be normal or only mildly enlarged.

Differential diagnosis

1. Hypertrophic cardiomyopathy—This can present with a history very similar to that of aortic stenosis. By contrast, the carotid pulse is normal or jerky

rather than slow rising. The diagnosis is confirmed by echocardiography, which reveals a normal aortic valve and shows characteristic ventricular features.

2. Congestive cardiomyopathy—Patients with long-standing untreated aortic stenosis can present with severe breathlessness, a large heart on radiography, a small volume pulse with a normal upstroke, a third heart sound, and pansystolic murmur due to papillary muscle dysfunction. These features can all be found in congestive cardiomyopathy. In endstage aortic stenosis, the echocardiogram shows a calcified valve with a significant (more than 35 mmHg) pressure drop across it: in congestive cardiomyopathy it shows a dilated and poorly contractile ventricle, but the aortic valve is normal.

3. Fixed subaortic stenosis—This is usually discovered in asymptomatic children and young adults in whom a systolic murmur is detected on routine examination.

An ejection click is absent, and a short early diastolic murmur usually heard. There is clinical and ECG evidence of left ventricular disease, which may be severe. The two-dimensional echocardiogram usually demonstrates the site and type of obstruction.

Prognosis. AS may progress slowly or quickly and thus requires regular follow-up to detect progression, particularly in sedentary elderly patients. In such patients, flow may become significantly compromised without triggering symptoms.

Overall, about 3 to 6% of asymptomatic patients with normal systolic function develop symptoms or LV ejection fraction depression every year. However, surgery is usually delayed until symptoms develop because the risk of surgery outweighs the survival benefit in asymptomatic patients. Surgery should not be delayed once symptoms develop. Mean survival in untreated symptomatic patients is about 2 to 3 yr. Aortic valve replacement relieves symptoms and improves survival. Risk with surgery increases for patients who require simultaneous coronary artery bypass graft (CABG) and for those with depressed systolic LV function. About 50% of deaths occur suddenly. While awaiting surgery, patients with severe AS should be advised to restrict physical exertion.

Treatment

- **Aortic valve replacement**
- **Balloon valvuloplasty**

Medical. Generally, any medical therapy has relatively poor effect in treating aortic stenosis. It is useful, however, in management of concomitant conditions that correlate with aortic stenosis:

- Any angina is generally treated with *short-acting nitrovasodilators, beta-blockers and/or calcium blockers.*

• Any ***hypertension*** is treated aggressively, but caution must be taken in administering ***beta-blockers***.

Any ***heart failure*** is generally treated with ***digoxin***, ***diuretics***, ***nitrovasodilators*** and if not contraindicated, cautious inpatient administration of ***ACE inhibitors***.

Since calcific aortic stenosis shares many pathological features and risk factors with ***atherosclerosis***, and since atherosclerosis may be prevented and/or reversed by cholesterol lowering, there has been interest in attempting to modify the course of calcific aortic stenosis by cholesterol lowering with ***statin*** drugs. Although a number of small, observational studies demonstrated an association between lowered cholesterol and decreased progression, and even regression, of calcific aortic stenosis.

Aortic regurgitation. Aortic regurgitation (AR), is the leaking of the ***aortic valve*** of the ***heart*** that causes blood to flow in the reverse direction during ventricular ***diastole***, from the ***aorta*** into the ***left ventricle***.

Etiology

• Acute aortic regurgitation.

- Rheumatic.
- Infective endocarditis.
- Ruptured sinus of Valsalva.
- Trauma, prosthetic valve surgery.
- Aortic dissection, laceration of the aorta.

• Chronic aortic regurgitation.

- Rheumatic.
- Syphilis.
- Aortitis (ie, ***Takayasu disease***).
- ***Marfan syndrome***.
- ***Osteogenesis imperfecta***.
- Bicuspid aortic valve, defect of the interventricular septum or sinus

of Valsalva.

- ***Ankylosing spondylitis***.
- ***Reiter syndrome***.
- Rheumatoid arthritis.
- Systemic lupus erythematosus.
- Hypertension.
- Infective endocarditis.

Clinical features

Symptoms. Patients with aortic regurgitation remain asymptomatic for many years. When symptoms develop, they are those of left ventricular disease, with:

- limitation of exercise tolerance by breathlessness or chest pain the most prominent one. Less commonly,

- the presenting symptom may be nocturnal dyspnoea, or an attack of acute pulmonary oedema.

- Retrosternal pain, aggravated by exertion, may develop in patients with aneurysms of the ascending aorta in whom the coronary arteries are normal. This seems to originate from the aortic root itself. Aortic dissection may also cause severe central chest pain.

Physical signs

- The hallmark of aortic regurgitation/insufficiency is a high-pitched decrescendo diastolic murmur at the left sternal border after the second heart sound.

- Acute aortic regurgitation

- Patients who have CHF or shock associated with severe aortic regurgitation often appear gravely ill.

- Tachycardia.

- Peripheral vasoconstriction.

- Cyanosis.

- Pulmonary edema.

- Arterial pulsus alternans; normal left ventricular impulse.

- Early diastolic murmur (lower pitched and shorter than in chronic aortic regurgitation) may be present. An Austin-Flint murmur, which is caused by the regurgitant flow causing vibration of the mitral apparatus, is lower pitched and short in duration. The decrescendo diastolic murmur is heard best with the patient leaning forward in full expiration in a quiet room. It is the cardiac murmur most commonly missed.

- A murmur at the right sternal border is associated more often with dissection than any other cause of aortic regurgitation.

- Chronic aortic regurgitation.

- All auscultatory phenomena indicate vasodilatation of peripheral circulation.

- Hyperdynamic apical impulse displaced laterally and inferiorly may be associated with an ejection click.

- Decrescendo diastolic murmur is heard best while the patient is leaning forward on deep expiration.

- Apical middiastolic rumble.

- Austin-Flint murmur.

- Pulsus bisferiens; increased pulse pressure; visible, forceful, and bounding peripheral pulses (water hammer).

- Corrigan pulse - Quickly collapsing pulses. Visible arterial pulsation in the neck.

- Musset sign - Bobbing of the head.
- Quincke sign - Capillary pulsations of the nail bed.
- Muller sign - Pulsations of the uvula.
- Hill sign - Systolic pressure in lower extremity greater than systolic pressure in upper extremity by at least 100 mm Hg.
- Traube sign - Loud systolic sound over femoral arteries.
- Duroziez sign - Systolic-diastolic murmur produced by compression of femoral artery with a stethoscope.

Investigations

• **transthoracic echocardiography**, which can provide two-dimensional views of the regurgitant jet, allow measurement of velocity using Doppler, and estimate jet volume.

The findings in severe aortic regurgitation, based on the 2006 American College of Cardiology/American Heart Association guidelines include:

- An AI color jet width > 65 percent of the left ventricular outflow tract (LVOT) diameter (may not be true if the jet is eccentric)
- Doppler vena contracta width > 0.6cm
- The pressure half-time of the regurgitant jet is < 250 msec
- Early termination of the mitral inflow (due to increase in LV pressure due to the AI.)

- Holodiastolic flow reversal in the descending aorta.

- Regurgitant volume > 60 ml

- Regurgitant fraction > 50 percent

- Regurgitant orifice area > 0.3 cm²

- Increased left ventricular size

In acute aortic regurgitation, echocardiography may show early closure of the mitral valve.

- **Chest X-ray** can assist in making the diagnosis, showing left ventricular hypertrophy and dilated aorta.

- **ECG** typically indicates left ventricular hypertrophy.

- **Cardiac chamber catheterization** assists in assessing the severity of regurgitation and any left ventricular dysfunction.

Differential diagnosis

Aortic regurgitation should also be distinguished from other causes of aortic run-off:

1. persistent ductus arteriosus;
2. ruptured sinus of Valsalva aneurysm; and
3. coronary arteriovenous fistula.

Treatment. Aortic insufficiency can be treated either medically or surgically, depending on the acuteness of presentation, the symptoms and signs associated with the disease process, and the degree of left ventricular dysfunction.

Surgical treatment is controversial in asymptomatic patients, however has been recommended if the ejection fraction falls to 50% or below, in the face of progressive and severe left ventricular dilatation, or with symptoms or abnormal response to exercise testing. For both groups of patients, surgery before the development of worsening ejection fraction/LV dilatation, is expected to reduce the risk of sudden death, and is associated with lower peri-operative mortality. Also, surgery is optimally performed immediately in acute cases.

Medical treatment. Medical therapy of chronic aortic insufficiency that is stable and asymptomatic involves the use of *vasodilators*. Small trials have shown a short term benefit in the use of *ACE inhibitors or angiotensin II receptor antagonists, nifedipine, and hydralazine* in improving left ventricular wall stress, ejection fraction, and mass. The use of these vasodilators is only indicated in individuals who suffer from *hypertension* in addition to AI. The goal in using these pharmacologic agents is to decrease the *afterload* so that the left ventricle is somewhat spared. The regurgitant fraction may not change significantly, since the gradient between the aortic and left ventricular pressures is usually fairly low at the initiation of treatment.

Other rather conservative medical treatments for stable and asymptomatic cases include *low sodium diet, diuretics, digoxin, calcium blockers and avoiding very strenuous activity*.

In addition, *endocarditis prophylaxis* is indicated before dental, *gastrointestinal* or *genitourinary* procedures.

In mild to moderate cases, echocardiography and cardiac stress test should be followed up every 1-2 years. In severe moderate/severe cases, echocardiography with cardiac stress test and/or *isotope perfusion imaging* should be performed every 3-6 months.

Surgical treatment. The surgical treatment of choice at this time is an *aortic valve replacement*. This is currently an open-heart procedure, requiring the individual to be placed on *cardiopulmonary bypass*.

Tricuspid regurgitation (TR). Tricuspid regurgitation (TR) is insufficiency of the tricuspid valve causing blood flow from the right ventricle to the right atrium during systole. The most common cause is dilation of the right ventricle. Symptoms and signs are usually absent, but severe TR can cause neck pulsations, a holosystolic murmur, and right ventricular-induced heart failure or atrial fibrillation. Diagnosis is by physical examination and echocardiography. TR is usually benign and does not require treatment, but some patients require annuloplasty or valve repair or replacement.

Etiology. TR is most commonly caused by dilation of the right ventricle (RV) with malfunction of a normal valve, as occurs in pulmonary hypertension, RV dysfunction-induced heart failure (HF), and pulmonary outflow tract obstruction. TR results less commonly from infective endocarditis in IV drug

abusers, carcinoid syndrome, chest or abdominal injury, rheumatic fever, idiopathic myxomatous degeneration, ischemic papillary muscle dysfunction, congenital defects (eg, cleft tricuspid valve, endocardial cushion defects), Ebstein's anomaly (downward displacement of a distorted tricuspid cusp into the RV), Marfan syndrome, and use of certain drugs (eg, ergotamine, fenfluramine, phentermine). Long-standing severe TR may lead to RV dysfunction–induced HF and atrial fibrillation (AF).

Symptoms and Signs. TR usually causes no symptoms, but some patients experience neck pulsations due to elevated jugular pressures. Acute or severe TR may cause symptoms of RV dysfunction–induced HF. Patients may also develop symptoms of AF or atrial flutter.

Pedal edema or ascites can occur in severe TR.

The only visible sign of moderate to severe TR is jugular venous distention, with a prominent merged *c-v* wave and a steep *y* descent. In severe TR, a right jugular venous thrill may be palpable, as may systolic hepatic pulsation and an RV impulse at the left lower sternal border. On auscultation, the 1st heart sound (S_1) may be normal or barely audible if a TR murmur is present; the 2nd heart sound (S_2) may be split (with a loud pulmonic component [P_2] in pulmonary hypertension) or single because of prompt pulmonic valve closing with merger of P_2 and the aortic component (A_2). An RV 3rd heart sound (S_3) may be audible near the sternum with RV dysfunction–induced HF.

The murmur of TR is a holosystolic murmur heard best at the left middle or lower sternal border or at the epigastrium with the bell of the stethoscope when the patient is sitting upright or standing. The murmur may be high-pitched if TR is trivial and due to pulmonary hypertension, or it may be medium-pitched if TR is severe and has other causes. Sometimes the murmur is not present at all and the diagnosis is best made by the appearance of the jugular venous wave pattern and the presence of hepatic systolic pulsations. The murmur varies with respiration, becoming louder with inspiration (Carvallo's sign).

Diagnosis

•Echocardiography

Mild TR is most often detected on echocardiography done for other reasons. More moderate or severe TR may be suggested by history and physical examination and confirmed by Doppler echocardiography. An ECG and chest x-ray are also often obtained.

•ECG is usually normal but, in advanced cases, may show tall peaked P waves caused by right atrial enlargement, a tall R or QR wave in V_1 characteristic of RV hypertrophy, or AF.

•Chest x-ray is usually normal but, in advanced cases with RV hypertrophy or RV dysfunction–induced HF, may show an enlarged superior vena

cava, an enlarged right atrial or RV silhouette (behind the upper sternum in the lateral projection), or pleural effusion.

Cardiac catheterization is rarely indicated for evaluation of TR. When catheterization is indicated (eg, to evaluate coronary anatomy), findings include a prominent right atrial *c-v* wave during ventricular systole.

Prognosis. Few reliable data about prognosis exist because so few patients develop severe TR in isolation.

Treatment

- Treatment of cause
- Sometimes valve repair or replacement

TR is usually well tolerated and often does not require surgical treatment. Medical treatment of causes (eg, HF, endocarditis) is indicated. The tricuspid valve may be repaired during surgery for left-sided heart lesions, such as mitral stenosis or regurgitation. Surgery may also be indicated for TR alone when RV impairment or hepatic cirrhosis threatens.

If endocarditis has damaged the tricuspid valve and cannot be cured with antibiotics, the valve may be totally excised and not replaced until 6 to 9 mo later; this procedure is well tolerated.

Tricuspid stenosis (TS). Tricuspid stenosis (TS) is narrowing of the tricuspid orifice that obstructs blood flow from the right atrium to the right ventricle. Almost all cases result from rheumatic fever. Symptoms include a fluttering discomfort in the neck, fatigue, cold skin, and right upper quadrant abdominal discomfort. Jugular pulsations are prominent, and a presystolic murmur is often heard at the left sternal edge in the 4th intercostal space and is increased during inspiration. Diagnosis is by echocardiography. TS is usually benign, requiring no specific treatment, but symptomatic patients may benefit from surgery.

TS is almost always due to rheumatic fever; tricuspid regurgitation is almost always also present, as is a mitral valve disorder (usually mitral stenosis). Rare causes of TS include SLE, right atrial (RA) myxoma, congenital malformations, and metastatic tumors. The RA becomes hypertrophied and distended, and sequelae of right heart disease–induced heart failure develop but without right ventricular (RV) dysfunction; the RV remains underfilled and small. Uncommonly, atrial fibrillation occurs.

Symptoms and Signs. The only symptoms of severe TS are fluttering discomfort in the neck (due to giant *a* waves in the jugular pulse), fatigue and cold skin (due to low cardiac output), and right upper quadrant abdominal discomfort (due to an enlarged liver).

The primary visible sign is a giant flickering *a* wave with gradual *y* descent in the jugular veins. Jugular venous distention may occur, increasing with inspiration (Kussmaul's sign). The face may become dusky and scalp veins may

dilate when the patient is recumbent (suffusion sign). Hepatic congestion and peripheral edema may occur.

On auscultation, TS may produce a soft opening snap and a mid-diastolic rumble with presystolic accentuation. The murmur becomes louder and longer with maneuvers that increase venous return (exercise, inspiration, leg-raising, Müller's maneuver) and softer and shorter with maneuvers that decrease venous return (standing, Valsalva maneuver).

Diagnosis

•Echocardiography

Diagnosis is suspected based on history and physical examination and confirmed by Doppler echocardiography showing a pressure gradient across the tricuspid valve. Two-dimensional echocardiography shows thickened leaflets with reduced movement and RA enlargement.

•**ECG** may show RA enlargement out of proportion to RV hypertrophy and tall, peaked P waves in inferior leads and V₁.

•**Chest x-ray** may show a dilated superior vena cava and RA enlargement, indicated by an enlarged right heart border. Liver enzymes are elevated because of passive hepatic congestion.

•**Cardiac catheterization** is rarely indicated for evaluation of TS. When catheterization is indicated (eg, to evaluate coronary anatomy), findings include elevated RA pressure with a slow fall in early diastole and a diastolic pressure gradient across the tricuspid valve.

Treatment

•Diuretics and aldosterone antagonists

•Rarely valve repair or replacement

Evidence to guide treatment is scarce. For all symptomatic patients, treatment should include a low-salt diet, diuretics, and aldosterone antagonists. Patients with hepatic congestion leading to cirrhosis or severe systemic venous congestion and effort limitation may benefit from interventions such as balloon valvotomy or valve repair or replacement. Comparative outcomes are unstudied.

6. TESTS AND ASSIGNMENTS FOR SELF-ASSESSMENT

FINAL LEVEL OF KNOWLEDGES

1. A 45-year-old woman has had worsening shortness of breath for 3 years. She now has to sleep sitting up on two pillows. She has had difficulty swallowing for the past year. She has no history of chest pain. She is afebrile. On physical examination palpation of the carotid arteries reveals a hypokinetic pulse. Palpation of the precordium reveals a right ventricular heave and the absence of the left ventricular impulse. Auscultatory findings include a loud S₁, most prominent is an early diastolic opening snap and decrescendo-crescendo rumbling diastolic murmur, heard best at the apex. A chest radiograph reveals a near-normal left ventricular size with a prominent left atrial border. An echocardiogram shows decreased opening of the mitral valve leaflets, and increased blood flow velocity during diastole. The trans-mitral gradient as measured by Doppler echocardiography is 10. Which of the following conditions is most likely to account for these findings?

A. Mitral valve stenosis.

D. Cardiomyopathy.

B. Aortic coarctation.

E. Essential hypertension.

C. Left renal artery stenosis.

2. A patient presents to the cardiology clinic with a new onset pansystolic murmur after a recent ST elevation anterior MI. Physical examination reveals a woman in no apparent distress with a heart rate of 110 bpm, blood pressure of 95/50 mm Hg, jugular venous pressure of 10 cm above right atrium, and bibasilar rales. Extremities are cool with 1+ edema. The echocardiogram reveals a ventricular septal rupture with left-to-right shunting and no valvular or paravalvular leak. Which of the following interventions has been associated with decreased mortality rates in stable patients with post-MI ventricular septal rupture?

A. Early surgical closure.

E. IV dopamine.

B. IABP.

F. IV fluid administration.

C. IV dopamine.

G. IV nitroprusside.

D. IV fluid administration.

3. A 45-year-old man is admitted to the intensive care unit with symptoms of congestive heart failure. He is addicted to heroin and cocaine and uses both drugs daily via injection. His blood cultures have yielded methicillin-sensitive *Staphylococcus aureus* in four of four bottles within 12 h. His vital signs show a blood pressure of 110/40 mmHg and a heart rate of 132 beats/min. There is a IV/VI diastolic murmur heard along the left sternal border. A schematic representation of the carotid pulsation is shown in the figure below. What is the most likely cause of the patient's murmur?

A. Aortic regurgitation.

B. Aortic stenosis.

C. Mitral stenosis.

D. Mitral regurgitation.

E. Tricuspid regurgitation.

4. A 50-year-old man has had progressive exertional dyspnea and chest pain for the past 2 months. Palpation of the carotid arteries reveals a diminished and delayed pulse. Palpation of the precordium reveals a dilated, sustained apical or left ventricular impulse with a presystolic impulse. On physical examination auscultation of the chest reveals an easily heard systolic, crescendo-decrescendo murmur is heard loudest at the upper right sternal border, and radiates to the carotid arteries bilaterally. An echocardiogram show a thickened, calcified aortic valve which opens poorly. Mean gradient on either side of the aortic valve is 50 mm Hg.

What is the most likely cause of the patient's condition?

A. Aortic stenosis.

B. Hypertrophic cardiomyopathy.

C. Mitral stenosis.

D. Pulmonic stenosis.

E. Ventricular fibrillation.

5. A 72-year-old male comes to the office with intermittent symptoms of dyspnea on exertion, palpitations, and cough occasionally productive of blood. On cardiac auscultation, a low-pitched diastolic rumbling murmur is faintly heard toward the apex. The origin of the patient's problem probably relates to

A. Rheumatic fever as a youth.

B. Long-standing hypertension.

C. Silent MI within the past year.

D. Congenital origin.

Навчальне видання

**Модуль 3. Внутрішня медицина.
Змістовний модуль №1.
Ведення хворих у кардіології.
Тема 15. Ведення пацієнта з шумом у серці**

*Методичні вказівки
для студентів та лікарів-інтернів*

Упорядники Бабак Олег Якович
 Фадєєнко Галина Дмитрівна
 Зайченко Ольга Євгенівна
 Шапошнікова Юлія Миколаївна
 Тверетінов Олександр Борисович

Відповідальний за випуск Бабак О.Я.

Комп'ютерний набір
Зайченко О.Є.

План 2012, поз. 24.
Підп. до друку 12.06.2012. Формат А5. Папір типогр. Різографія.
Умов. друк. арк. 2,25. Тираж 150 прим. Зам. № 12-2933.

**Редакційно-видавничий відділ
ХНМУ, пр. Ленина, 4, г. Харків, 61022
izdat@knhu.kharkov.ua**

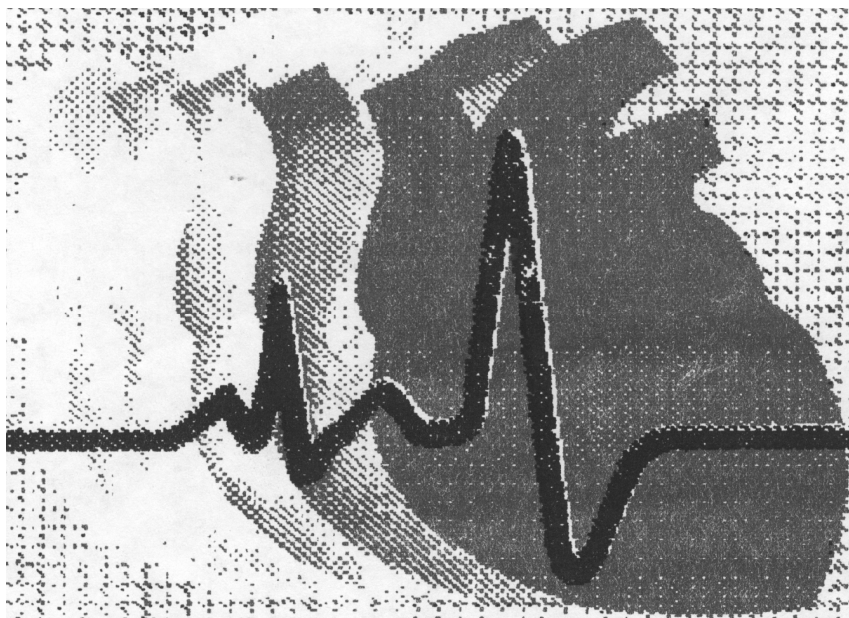
Свідоцтво про внесення суб'єкта видавничої справи до Державного реєстру видавництв, виготовників і розповсюджувачів видавничої продукції серії ДК № 3242 від 18.07.2008 р.

Module. Internal medicine.

**Meaning module №1. Keeping patients
in cardiological clinic.**

**Theme 15. Management of patient
with a heart murmur**

Guidelines for students



Модуль. Внутрішня медицина.

Змістовний модуль №1.

Ведення хворих у кардіології. Тема 15.

Ведення пацієнта з шумом у серці

*Методичні вказівки
для студентів та лікарів-інтернів*